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Polymyositis as presenting manifestation of gallbladder carcinoma: A case report

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ABSTRACT

INTRODUCTION: Inflammatory myositis as a paraneoplastic presentation of gallbladder cancer is an extremely rare event. In this paper we reported the first case of gallbladder carcinoma presented as polymyositis.
PRESENTATION OF CASE: A 68-year-old housewife presented with proximal muscles weakness, pain, significant decrease in force of proximal muscles, and globally decreased deep tendon reflexes. Laboratory studies revealed an anemia, increased acute phase reactants and increased serum creatine phosphokinase (CPK) levels. Electromyography (EMG) and nerve conduction velocity test (NCV) demonstrated mild myopathic changes. Muscle biopsy was suggestive for polymyositis. Corticosteroid therapy initiated and a meticulous search for probable underlying malignancy performed concurrently. Malignancy workup finally revealed a gallbladder tumor. Patient candidated for extended cholecystectomy. Pathologic evaluation of gallbladder tumor demonstrated a moderately differentiated carcinoma. Progressive improvement in clinical conditions and complete normalization of laboratory parameters occurred postoperatively. After 8 months of follow-up patient is still alive and in good state of health. There is no evidence of metastatic or local recurrence of tumor. Musculoskeletal complaints subsided completely.
DISCUSSION: Gallbladder carcinoma is a rare and usually aggressive malignancy. Its primary presentation by paraneoplastic syndromes especially in the form of paraneoplastic neurological syndromes is an extremely rare event. Some believes that increased association between inflammatory myopathy and malignancy is limited to the dermatomyositis; however, presentation of our patient as polymyositis is contrary to this. This is the first reported case of gallbladder cancer who presented with polymyositis.
CONCLUSION: Gallbladder cancer though rare, should be considered in patients with inflammatory myositis.

1. Introduction

Inflammatory myositis (including dermatomyositis/polymyositis) is an inflammatory myopathy which accompanied by muscle weakness as its hallmark symptom. This condition is associated with malignancies in a significant minority of cases. An association between malignancy and inflammatory myopathy has been suspected as early as 1916, when Stertz reported the simultaneous occurrences of PM and gastric carcinoma.[1] The close relationship between inflammatory myopathy and cancer is consistent with the concept that paraneoplastic processes linked to oncogenesis and autoimmunity contribute to the disease in a subset of these cases. However, the precise links between malignancy and inflammatory myopathy remain incompletely understood.[2,3]

This case report presented a gallbladder carcinoma manifested as polymyositis, which is to our knowledge the fourth known case in the world of this malignancy associated with inflammatory myopathy despite the previous three cases presented by dermatomyositis; this is the first reported case of gallbladder carcinoma which was associated with polymyositis.

2. Case presentation

A 68 year old housewife presented with proximal muscles weakness and pain since two weeks prior to admission. Patient had no previous history of similar problem and/or hospitalization due to any medical or surgical condition. On examination, there was a significant decrease in force of proximal muscles of both upper and lower extremities which was more prominent in upper limbs. Proximal muscle tenderness was also notable. However, distal muscles had a relatively normal force. Deep tendon reflexes were decreased globally. There was no other significant physical finding on examination.

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Fig. 1. Macroscopic appearance of the specimen (gallbladder tumor).

Initial laboratory studies indicated a normocytic hypochromic anemia associated with leukocytosis and mild thrombocytopenia. Serum CPK level was 22,250 IU/L (normal up to 170). Acute phase reactants including the erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) levels were also increased. Other abnormal findings included increased aspartate aminotransferase (AST), alanine aminotransferase (ALT), Lactate dehydrogenase (LDH) and triglyceride levels; and decreased total protein, albumin, calcium, High-density lipoprotein (HDL) and Low-density lipoprotein (LDL) cholesterol levels.

Thyroid function, renal function, bilirubin, alkaline phosphatase and immune markers including anti-nuclear antibody (ANA), anti double stranded deoxytribonucleic acid (Anti ds-DNA), anti-cyclic citrullinated peptide antibody (anti-CCP), and anti JO-1 antibody were all within normal limits. Serum viral hepatitis markers were also negative (Fig. 1).

EMG/NCV study demonstrated mild myopathic changes.

Patient underwent muscle biopsy which revealed inflammatory myopathy, which was reported most probably suggestive for polymyositis (Figs. 2 and 3).

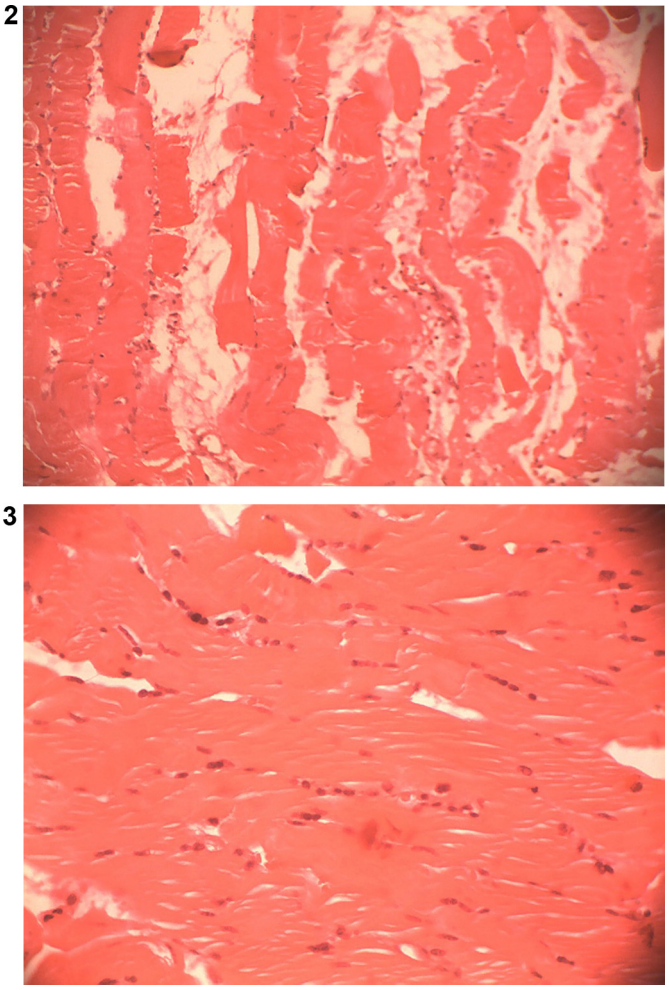
Corticosteroid therapy initiated with 60 mg Prednisolon (1 mg/kg) in divided daily doses and a meticulous search for probable underlying malignancy performed concurrently. Malignancy workup finally revealed a gallbladder tumor with increased level of Carcinoembryonic antigen (CEA) and normal level of cancer antigen (CA) 19-9 and CA 125.

The patient candidated for surgical intervention and extended cholecystectomy with abdominal sampling undertaken. Pathologic evaluation of gallbladder tumor demonstrated a moderately differentiated carcinoma which was at least in stage IIA (Figs. 4–8) and abdominal samples were negative for malignancy.

Corticosteroid therapy continued postoperatively for 10 days with pre-operative dose and then gradually corticosteroid decreased to 5 mg/day within 2 weeks and discharged home with this dose and prednisolone subsequently discontinued 1 month after discharge.

Patient responded well to the surgery and a progressive improvement in clinical condition and complete normalization of laboratory parameters observed. Upper extremity muscular force at discharge was 4/5 and force of lower limb was 5/5. It should be noted that, a common bile duct (CBD) stone complicated the post-operative recovery course of the patient, which managed endoscopically.

After 8 months of post-operative follow-up, patient is still alive and in good state of health. There is no evidence of metastatic or local recurrence of tumor and musculoskeletal complaints subsided completely.

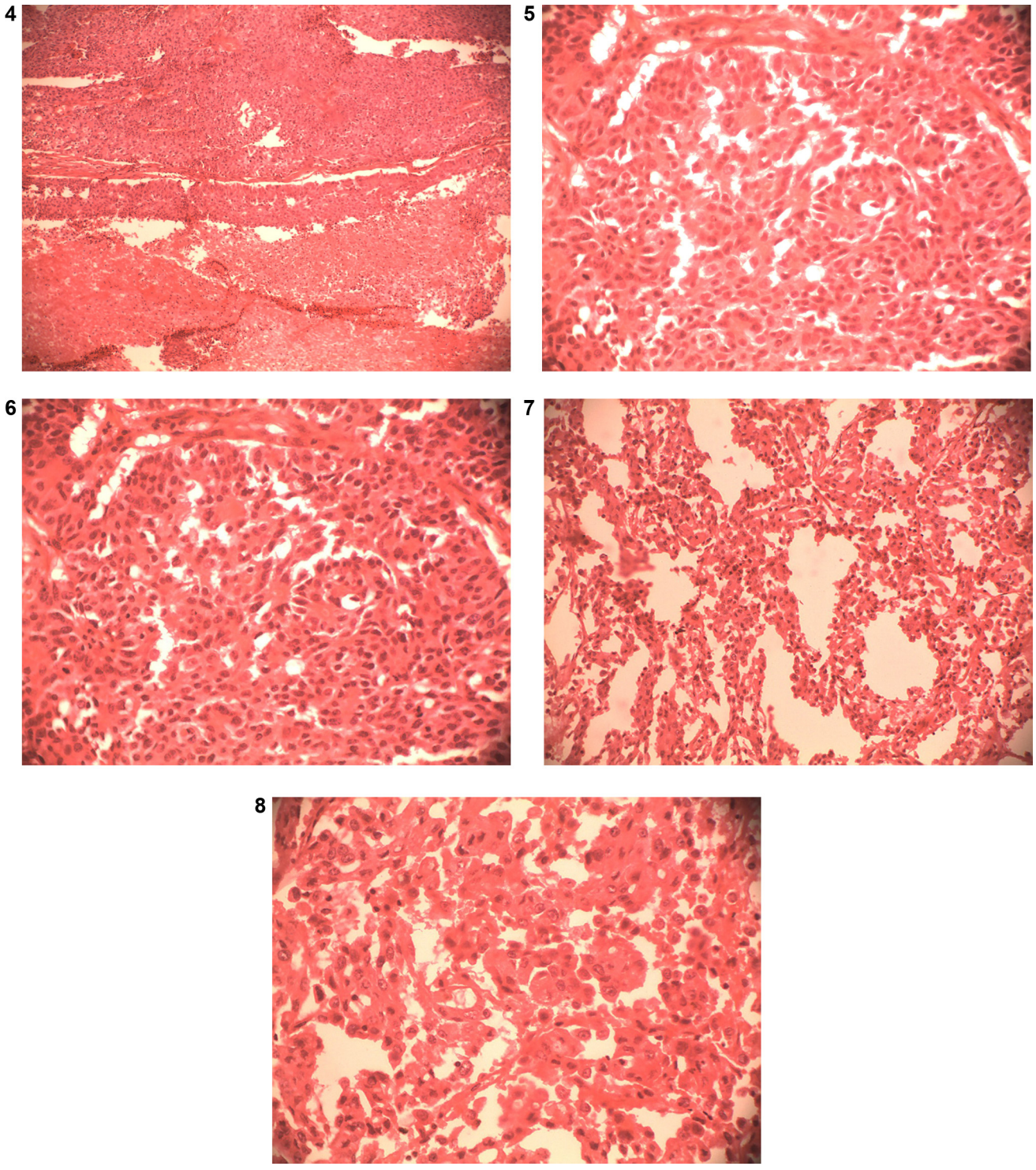


Figs. 2 and 3. Microscopic feature of polymiositis.

3. Discussion

Gallbladder carcinoma is a rare and usually aggressive malignancy that occurs predominantly in the elderly[4] and its primary presentation by paraneoplastic syndromes especially in the form of paraneoplastic neurological syndromes (PNS) is an extremely rare event. PNS are defined as remote cancer effects which are neither due to tumoral invasion or its metastasis nor due to ischemic, metabolic, infectious, nutritional or therapeutic consequences of the primary tumor.[5,6] Any level of central or peripheral nervous system including neuromuscular junction and muscle may be affected in PNS.[6] Inflammatory myopathies are well known PNSs and are most commonly associated with ovarian cancer, lung cancer, breast cancer, melanoma, colorectal cancer and non-Hodgkin's lymphoma.[6,7] Estimates of the associated risk of malignancy with inflammatory myopathies vary widely in different studies (between 6 and 60).[8]

Some believes that increased association between inflammatory myopathy and malignancy is limited to the dermatomyositis[7]; however, presentation of our patient as polymyositis is contrary to this. It should be noted that this is the first reported case of gallbladder cancer who presented with polymyositis. In all other cases of myopathic presentations of gallbladder cancer which include only 3 published reports, patients presented with dermatomyositis.[9–11]



Figs. 4–8. Microscopic appearance of gallbladder adenocarcinoma.

In conclusion, gallbladder cancer though rare, but should be considered in patients with both dermatomyositis and polymyositis.

Conflicts of interest

The authors declare no conflicts of interest on this work.

Ethical approval

This study was approved by the Research Committee of the Arak University of Medical Sciences. Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Babak Adli and Mohsen Pakzad collected and interpreted the patient's data. Mohammad Naeem Bangash and Mohsen Pakzad were major contributors in writing the manuscript. Babak Adli and Mohsen Pakzad and Siamak Rakei performed surgery. All authors read and approved the final manuscript.

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References

1. Stertz O. Polymyositis. *Klinische Wochenschrift (Berlin)* 1916;**53**:489.
2. Casciola-Rosen L, Nagaraju K, Plotz P, Wang K, Levine S, Gabrielson E, et al. Enhanced autoantigen expression in regenerating muscle cells in idiopathic inflammatory myopathy. *Journal of Experimental Medicine* 2005;**201**(February (4)):591–601.
3. Levine SM. Cancer and myositis: new insights into an old association. *Current Opinion in Rheumatology* 2006;**18**(November (6)):620–4.
4. Brunicard FC, Andersen DK, Billiar TR, Dunn DL, Hunter JG, Matthews JB, et al. *Schwartz's principles of surgery*. 9th ed. USA: McGraw-Hill; 2010.
5. de Beukelaar JW, Sillevis Smitt PA. Managing paraneoplastic neurological disorders. *Oncologist* 2006;**11**(March (3)):292–305.
6. Honnorat J, Antoine JC. Paraneoplastic neurological syndromes. *Orphanet Journal of Rare Diseases* 2007;**May (2)**:22.
7. Chawla J. Stepwise approach to myopathy in systemic disease. *Frontiers in Neurology* 2011;**2**(49), http://dx.doi.org/10.3389/fneur.2011.00049 [Epub ahead of print].
8. Sigurgeirsson B, Lindelof B, Edhag O, Allander E. Risk of cancer in patients with dermatomyositis or polymyositis A population-based study. *New England Journal of Medicine* 1992;**326**(6):363–7.
9. Kundu AK, Karmakar PS, Bera AB, Pal SK. Carcinoma of the gall bladder presenting as dermatomyositis. *Journal of the Association of Physicians of India* 2005;**March (53)**:219–22.
10. Yiannopoulos G, Ravazoula P, Meimaris N, Stavropoulos M, Andonopoulos AP. Dermatomyositis in a patient with adenocarcinoma of the gall bladder. *Annals of the Rheumatic Diseases* 2002;**61**(July (7)):663–4.
11. Narasimhaiah DA, Premkumar JA, Moses V, Chacko G. Carcinoma of gall bladder presenting as dermatomyositis. *Annals of Indian Academy of Neurology* 2011;**14**(January (1)):44–6.